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REHABILITATION IN PRACTICE

Long-term functional outcome of patients with longitudinal radial deficiency: cross-sectional evaluation of function, activity and participation

Irene Holtslag^{1,2}, Iris van Wijk^{1,3}, Hermien Hartog⁴, Aebele Mink van der Molen² & Corry van der Sluis⁴

¹Department of Rehabilitation Medicine, University Medical Centre of Utrecht, Utrecht, The Netherlands, ²Department of Plastic and Reconstructive Surgery, University Medical Centre of Utrecht, Utrecht, The Netherlands, ³Rehabilitation Center de Hoogstraat Utrecht, Utrecht, The Netherlands, and ⁴Department of Rehabilitation Medicine, University of Groningen, University Medical Center Groningen, Groningen, The Netherlands

Purpose: To evaluate all functional aspects of patients with longitudinal radial dysplasia and to clarify the relationship between body functions on the one hand and limitations in activity and participation on the other hand. **Methods:** Thirty-one arms of seventeen adult patients with longitudinal radial dysplasia were analysed. Body function was assessed by measuring grip and pinch strength and active range of motion (ROM) of the hand. Activities were measured using the "Sequential Occupational Dexterity Assessment", to measure perceived restrictions in participation the "Impact on Participation and Autonomy questionnaire" was used. Relationships between severity of dysplasia, body function, participation and activity were determined. **Results:** Patients with a severe type scored significantly lower in body function scores than patients with a mild form. Patients with limited active finger joint motion performed worse on activities. We found no significant differences in activity and participation between mild or severe types and found no correlation in participation scores. **Conclusion:** Although considerable restrictions in joint mobility and strength were revealed, little or no limitations on the activity and participation level were found. Limitations in body functions hardly influenced capacity on activity level and did not influence participation in societal roles.

Keywords: Upper limb deformity, patient participation, activities of daily living, longitudinal radial deficiency

Introduction

Longitudinal radial deficiency (LRD) is a congenital malformation characterised by partial or total absence of the radius.

Implications for Rehabilitation

- People with LRD learn to accomplish many of the everyday tasks without great difficulty and do not report a low quality of participation in major life activities.
- Professionals working in rehabilitation medicine should focus on activity and participation rather than on body structure or functions. Therapy focused solely on increasing joint motion or strength does not lead to further improvement.
- For parents it will be reassuring to know that children with severe radial deficiencies can satisfactorily fulfill social roles in later life.

The disorder has a prevalence of 1 in 30,000 to 100,000 live births [1]. Little is known about the aetiology. Between the fourth and seventh post-conception weeks the limbs develop. During this period, environmental, genetic or idiopathic factors may create specific defects [1,2].

With partial absence of the distal radius or total absence of the radius, the wrist lacks the normal support of the forearm resulting in an instable wrist with characteristic radial deviation of the ulna and carpus: often with (sub)luxation of the wrist [1–5]. Traditionally, classification of LRD is based on the extent of hypoplasia or aplasia of the radius defined by Bayne and Klug [4]. The congenital defect, however, is not limited to only the radius; almost invariably muscular and neurovascular structures at the radial side of the entire forearm and wrist are involved as well, possibly aggravating functional deficiencies. Upper extremity functional deficiencies seen in LRD are limited arm length, due to a

short arched forearm, diminished finger function and grip strength due to hypoplasia, aplasia or digital contractures, loss of pro- and supination, ulnar oriented hand function and palmar flexion of the wrist during grasping [1–5].

The objectives of surgical treatment of LRD are to correct the position of the wrist, to (re)construct thumb function and to create adequate length of the forearm. The goal of these procedures is to make a more functional and aesthetically acceptable arm and hand [5].

In the clinical evaluation of hand function, it has been practice to focus on limitations in body functions by measuring grip and pinch strength and joint mobility [6]. A more complex aspect of function is the execution of daily tasks or the fulfillment of an individual's social role, which is referred to as activity and participation by the World Health Organisation in the International Classification of Functioning, Disability and Health (ICF) [7]. For the patient, restrictions in activity or participation may be more important than impairments in grip and pinch strength or joint mobility [6,8,9].

However, to our knowledge there is no information on the relationship between the severity of LRD and its influence on participation and activity.

The goal of this study was to evaluate aspects of function, activity and participation of adult patients with LRD according to terms defined by the ICF. Furthermore, we aimed to clarify the relationship between body function on the one hand and limitations in activity and participation on the other hand in patients with LRD in the current Dutch Society.

Materials and methods

Participants

In this retrospective cross-sectional study patient records of the two participating hospitals, the University Medical Centres of Groningen and Utrecht and Rehabilitation Centre “de Hoogstraat” Utrecht, the Netherlands, were reviewed for patients with LRD. Patients aged 18 years and older at the time of the study with LRD (Bayne and Klug types 1, 2, 3, or 4), surgically or conservatively treated, were included. Medical records of participating patients were reviewed for medical information and treatment history. The patients were asked to attend an outpatient clinic, where the first and third author performed physical examination and measurements, as outlined below. In addition the patients were asked to complete a questionnaire. Our study followed principles as described in the declaration of Helsinki. The local medical ethics committee of rehabilitation centre “de Hoogstraat” approved the study, but no formal approval was obtained, since measurement of range of motion (ROM) and strength is part of normal care and for filling in questionnaires no further ethical approval was needed. All patients signed a written informed consent.

Measurement instruments

Body function and structure

Power grip was measured using a digital Jamar dynamometer with the handlebar in position 2 (E-link, Biometrics). Tip-to-tip strength was measured using a digital pinch grip

dynamometer (E-link, Biometrics) and was defined as the grip between the tip of the thumb, or the tip of the middle or ring finger if absent, and the index finger. During the measurements participants were seated in a standard position; the elbows flexed in a 90-degree position without support of the table. The mean of three maximum voluntary contractions was recorded.

The active ROM of all joints of the hand was measured using a digital goniometer (E-link, Biometrics). The active ROM of the interphalangeal joints was classified according to the original Strickland-Glogovac method [10,11]: the active interphalangeal joint flexion minus the total interphalangeal joint extension divided by the normal average interphalangeal joint motion $\times 100$. The result is considered a percentage of normal and according to Strickland's criteria a total active ROM of the fingers was classified as excellent (220–260° equalizes 85–100% of normal ROM), good (180–219°: 70–84%), fair (130–179°: 50–69%) or poor (<130°: <50%). The active ROM of the thumb was calculated separately according to Strickland's criteria for ROM of the thumb (excellent; 119–140°: 85–100%, good; 98–118°: 70–84%, fair; 70–97°: 50–69% and poor; <70°: <50%). In case of pollicisation (surgically creating a thumb) we used the criteria for fingers to calculate the ROM of the pollicised finger. In this study, pollicisation was only done using the index finger.

Activities

Activities were measured using the Sequential Occupational Dexterity Assessment (SODA), an observational measurement instrument that provides a performance score for 12 standardized tasks of common daily activities [12]. The SODA has good psychometric properties [13].

For each task a prescribed functional grip is described. The patient performs the standardised tasks under controlled conditions and the trained researcher rates the performance. A first score is obtained for the ability of a patient to perform the tasks in the standardised way (4 points), in a different way (1 point), or the inability to perform the activity (0 points). A second score is derived from the effort it takes to perform a task, scored by the patient (much (0 points), little (1 point) or no effort (2 points)). A higher score represents a better performance and less effort, with a maximum score of 108 points. Six tasks require bimanual function, whereby each hand is allocated a single score. Each task is provided twice, left and right hand, to score each hand independently.

Participation

To measure perceived restrictions in participation in daily life roles, patients were asked to complete the Impact on Participation and Autonomy questionnaire (IPA). Thereby focussing on only the radial dysplasia [14]. The questionnaire consists of five domains: autonomy indoors (7 items, e.g. getting around at home, engaging in self-care activities), family role (7 items, e.g. housekeeping, economic self-sufficiency), autonomy outdoors (5 items, e.g. visiting friends, spending leisure time in a way the participant decides upon), social relationships (6 items, e.g. quality of relationships, receiving respect), and work and educational opportunities (6 items, e.g. performing the job

or obtaining the education of one's choice). Restrictions in participation and perceived problems are reflected in two separate scores with a range per item of 1–5 and 0–2, respectively. A higher score represents greater restrictions in participation or worse problems. A problem is severe when score 2 is given. The IPA is valid, reliable and responsive to change [15,16].

Subjective information

We enquired about satisfaction with the appearance of the affected limb at home and in public, influence of the disorder on mood and social life and the patient's opinion on surgical interventions in the past or in the future regarding function and aesthetics, using for each item a 100 mm Visual Analogue Scale (VAS). In total, we used eight VAS scores, whereby a 0 reflected the worst situation; a VAS score of 100 reflected the best situation, according to the patient.

Statistical methods

For reasons of analysis, the type of radial deficiency was dichotomized. Types 1 and 2 Bayne and Klug radial deficiency were classified as mild, and types 3 and 4 as severe.

As strength depends on gender, age and hand dominance, percentages of normal values of grip and pinch strength adjusted for gender, age and side were used in the analyses [17]. Strength and Strickland's ROM scores represent a single arm whereas SODA, IPA and IPA problem perception scores represent a person. In patients with bilateral defects, we used the SODA score of the most severely affected arm to relate to IPA and VAS scores under the assumption that the most severe type of LRD determines the degree of functional limitation in a person [18].

Mean function, activity and participation scores were calculated in total and per type of LRD. Variables were compared using the Mann–Whitney *U*-test, because numbers were low. To analyse the relationship between function on the one hand and activity and participation on the other hand correlations were calculated and expressed in Pearson's *r*. A *p* level of <0.05 was considered to be statistically significant.

Results

Patients

Twenty-three patients were identified. Seventeen patients with in total 31 affected arms participated in the study (Table I). All patients were self-sufficient besides one, she received a government allowance and has her own household. Of the six non-participants, four did not participate for personal reasons, one refused due to traffic distance and one had died.

Twelve patients (71%) had an associated syndrome: VACTERL association (vertebral anomalies, anal atresia, cardiac anomalies, tracheoesophageal fistula, renal anomalies, radial anomalies, lung anomalies, *n* = 4, 33%), Holt–Oram syndrome (*n* = 4, 33%), TAR syndrome (Thrombocytopenia with absent radius, *n* = 2, 17%), Klippel Feil syndrome (*n* = 1, 8%) and Fanconi Anemia (*n* = 1, 8%). Nineteen limbs of twelve patients had been treated operatively. Due to the retrospective design of this study, the actual reason for conservative or operative treatment was not clear.

Table I. Demographics of the study population.

	Frequency	%
Number of patients	17	
Gender male/female	9/8	53/47
Age (in years) median (IQR)	22 (11)	
<i>Occupation</i>		
Working	4	23
Student	8	47
Retired	3	18
Unknown	2	12
<i>Type of radial deficiency (Bayne and Klug)</i>		
Mild (1&2)	12	39
Severe (3&4)	19	61
Operated	19*	61
<i>Operation</i>		
Centralisation	12	63
Pollicisation	6	32
Other	3	16

IQR, interquartile range.

*Two patients had more than one type of surgical treatment.

Body function and structure

In 10 patients with severe type LRD, the thumb was absent. All patients, with 31 affected limbs, scored very low in power grip en tip-to-tip strength, 24.1% and 18.9% of norm values, respectively (Table II). Patients with severe LRD scored significantly lower than patients with mild LRD in ROM of fingers, power grip strength and pinch grip strength. We could not draw any conclusions about the effect of pollicisation, because only three patients underwent (bilateral) pollicisation all with a different type of LRD.

Activity and participation

We found no significant differences in activity and participation between patients with mild or severe type of LRD. SODA scores, total IPA scores and problem perception IPA scores did not differ significantly between both LRD groups (Table III).

Per factor IPA Participation scores were good. Autonomy indoor average score was 0.2 (median score 0.1), autonomy outdoor, family role, social relationships and work scored 0.5 (0.2), 0.6 (0.4), 0.6 (0.6), and 0.5 (0.7) respectively.

In the factors mobility, social and work for problem perception IPA, three different patients experienced severe problems. This was due to the lack of a driving license (mobility and social), and not being able to work in the construction industry as a forklift driver.

Five patients were students and could not completely fill in the IPA problem perception work and education questions due to lack of work experience.

Subjective information

VAS scores indicated that the radial deficiency has very little influence on patients' mood and social contacts (median VAS 99 (IQR = 30) and 90 (IQR = 40), respectively). Patients were moderately satisfied with the appearance of their affected limb at home and in public (median VAS score 60 (IQR = 44) and 67 (IQR = 41), respectively).

Table II. ROM, grip and pinch strength scores.

	Total		Type LRD			Mann-Whitney U-test	
	Median (IQR)	n	1&2 Median (IQR)	n	3&4 Median (IQR)	n	p value
ROM thumb	58.8 (39.2)	21	61.4 (32.1)	12	58.0 (67.0)	9	0.943
Strickland classification	Fair		Fair		Fair		
ROM fingers	65.6 (74.2)	31	91.5 (11.3)	12	33.4 (45.6)	19	0.003
Strickland classification	Fair		Excellent		Poor		
Power grip strength (%)	24.1 (24.3)	31	38.4 (17.1)	12	18.7 (12.9)	19	0.007
Pinch grip strength (%)	18.9 (30.9)	31	38.4 (48.7)	12	11.8 (19.2)	19	0.001

IQR, interquartile range; LRD, longitudinal radial deficiency; n, absolute number; P, significance; ROM, range of motion.

Patients scored very low on their expectations of improvement to the appearance of their limb by surgery (median VAS 25, IQR = 59). They seemed to be more confident in improvement of function by surgery (median VAS 73, IQR = 35). No significant differences were found in any of the VAS scores between patients with mild and severe LRD.

Correlation between function, activity and participation

Regarding the relation between function and activity, it was found that only the ROM of the fingers correlated significantly with SODA scores (Table V). Patients with greater active ROM had better performance in the SODA tasks.

Regarding the relation between function and participation none of the body function variables correlated significantly with total IPA score or IPA problem perception scores.

Operatively vs. conservatively treated patients

As centralisation is the major procedure mainly performed in LRD patients, a comparison is presented of outcomes on patients with severe LRD (Bayne and Klug types 3 and 4), who were treated either operatively by centralisation or conservatively (Table V). Since the thumb was present in only four patients Strickland thumb scores are not presented.

SODA and total IPA scores were relatively equal in the groups. Patients with unilateral LRD seem to experience more problems than patients with bilateral LRD although there is a great difference between minimum and maximum scores in the bilateral severe LRD patients who underwent a centralisation.

Furthermore, operatively treated patients were convinced that surgery improved their appearance and function (VAS 100 (IQR = 70) and VAS 100 (IQR = 72), respectively). Although VAS scores for the expectation that an operative treatment in the future might improve appearance or function were lower (VAS 41 (± 35) and VAS 65 (± 28), respectively). Conservatively treated patients also had low scores in their expectations that surgery would improve their appearance or function of their limb in the future (VAS 25 (± 43), VAS 65 (± 22), respectively).

Discussion

In this study adults with LRD were presented and also the relationship between three levels of the International Classification of Functioning were studied, namely; body

Table III. SODA, IPA, IPA problem perception scores.

	Total (n = 17) Median (IQR)	Type LRD		Mann-Whitney U-test p value
		1&2 (n = 4) Median (IQR)	3&4 (n = 13) Median (IQR)	
SODA	79.0 (9.5)	79.0 (11)	78.0 (15)	0.427
Total IPA	2.4 (2.5)	2.9 (3.4)	2.0 (2.5)	0.955
IPA problem perception	1.0 (3.0)	0.5 (1)	1.0 (4)	0.126

IPA, Impact on Autonomy and Participation Questionnaire; IQR, interquartile range; LRD, longitudinal radial deficiency; n, absolute number; P, significance; SODA, Sequential Occupational Dexterity Assessment.

function and structure, activity and participation. Although considerable restrictions in joint mobility and strength were revealed, surprisingly little or no limitations on the activity and participation level were found. Furthermore, there appeared to be hardly any relation between body functions on the one hand and activities and participation on the other hand.

Body function

The reduced active ROM and strength in patients with LRD has been described formerly in literature, especially in children [6,9]. It is also known that in severe LRD less ROM and strength is found compared to milder forms of LRD [2,6,9,19,20]. From our results it became clear that the impairments in body functions do not imply that adult LRD patients feel restricted in activities or participation, which is important information for parents of children with recently diagnosed LRD, especially severe LRD. These parents should also be informed that a limited ROM of the finger joints and limited strength will still be present, as the child grows older, with or without surgery. Furthermore, this knowledge is useful for professionals working in the field of rehabilitation medicine, they should focus on activity and participation rather than on body structure or functions when therapy focused on increasing joint motion or strength is not leading to further improvements.

Activity

The SODA was chosen to clarify into restrictions of activities in LRD patients. We realize that this measurement instrument is not validated for this specific population. However, no further suitable instrument was available. The SODA has been mainly used in patients with rheumatoid

arthritis (RA). In literature, SODA scores for RA patients vary between 60 and 88 [21,22]. Our results (median SODA score 79) indicate that our patients seem to experience minor to moderate restrictions in performing their activities. As the tasks of the SODA had to be performed in a prescribed manner, even better scores might have been derived if our patients had been allowed to perform the test in their own (adapted) way. During the tests patients reported that they had difficulty with some of the activities in the SODA test where tip-to-tip handle or tip-to-tip strength and/or supination of the forearm was needed, such as buttoning a shirt, squeezing a toothpaste tube, cutting meat and carrying heavy objects. These restrictions are comparable to results from previous reports from Buffart et al. [9] and Bora et al. [20]. All patients adapted well to their daily activities, although some used aids such as special cutlery or adaptations to trousers or shirts to be able to use buttons, like snap fasteners. Others avoided certain tasks, such as wearing shirts with buttons. To get a valid and reliable insight into the restrictions LRD patients encounter, we recommend a specific test to be designed for this group. Only then can results of different studies be compared or pooled. The latter is relevant since most studies deal with only small study populations like ours [6,9,19,20].

Table IV. Correlations pearson correlation coefficients.

	SODA <i>r</i> (<i>p</i> value)	IPA total <i>r</i> (<i>p</i> value)	IPA problem perception <i>r</i> (<i>p</i> value)
	<i>n</i> = 17		
ROM thumb	-0.28 (0.47)	0.30 (0.43)	0.02 (0.96)
ROM fingers	0.56 (0.02)	-0.13 (0.62)	-0.38 (0.14)
Power grip strength	0.09 (0.73)	0.06 (0.81)	-0.20 (0.45)
Pinch grip strength	0.45 (0.07)*	-0.04 (0.89)	-0.25 (0.34)

*For SODA pinch grip strength *n* = 14. Bold value is significant at the 0.01 level. IPA, Impact on Autonomy and Participation Questionnaire; *n*, absolute number; *r*, Pearson's correlation coefficient; ROM: range of motion; SODA, Sequential Occupational Dexterity Assessment.

Participation

Patients did not experience great restrictions in participation (median IPA total 2.4) compared to patients with RA (IPA total 2.5–3.6), neuromuscular diseases like post-polio syndrome, hereditary motor and sensory neuropathy (HMSN), limb girdle dystrophy or spinal muscular atrophy (SMA): IPA total 2.2 to 3.1. In patients with leprosy hand impairment contributed only minor in participation restrictions, perceived restrictions were mainly caused by eye impairment [23].

The proportion of patients perceiving severe problems in these groups was much higher than in our study group as we found a maximum of one person per subscale who experienced severe problems (6%) against 12 to 43% per subscale found by Cardol et al. [16]. This score is also lower than that reported for people with impairments due to leprosy. They experienced up to 21% severe problems in participation [23].

The mild restrictions in participation may be explained by the fact that LRD is a congenital disorder, which enables people to cope and develop adequate strategies to live their lives despite the disorder, and the IPA measures subjectively instead of objectively. [24] That may also explain why hardly any differences in activity and participation level between mild and severe LRD types was found. Notable is the variation in the scores, especially in IPA problem perception: some patients with mild LRD experienced severe problems, whereas others with severe LRD did not experience any problems. Previously, Cardol et al. [16] stated that the impact of illness on participation is individually specific and emotional distress is an important factor intermediating between disease and outcomes in terms of restrictions in participation. In patients with RA participation restrictions these are mainly explained by diminished vitality and mental health, which indicate that comprehensive scales of health covering psychological aspects like depression, wellbeing and mood have an extensive influence on participation in daily life [8].

Subjective information

From the subjective information derived with VAS-scores, it appeared that patients have accepted their deficiency and

Table V. Operatively and conservatively-treated patients.

	Severe LRD unilateral		Severe LRD bilateral		
	Centralisation		Centralisation		
	No	Yes	No	Unilateral	Bilateral
	Median (min-max) (<i>n</i> = 2)	Median (min-max) (<i>n</i> = 3)	Median (min-max) (<i>n</i> = 4)	Median (min-max) (<i>n</i> = 2) [‡]	Median (min-max) (<i>n</i> = 8) [‡]
ROM fingers	62.6 (20.3–104.8)	33.4 (15.3–52.0)	30.5 (15.4–47.1)	83.15 (65.6–100.7)	23.4 (8.1–92.4)
Strickland classification	Fair	Poor	Poor	Good	Poor
Power grip strength (%)	16.1 (13.8–18.4)	11.3 (0.0–37.1)	16.0 (5.9–24.7)	27.95 (23.3–32.6)	16.7 (7.8–51.2)
Pinch grip strength (%)	6.9 (0.0–13.8) (<i>n</i> = 2)	7.6 (0.0–26.7) (<i>n</i> = 3)	4.8 (0.0–37.8) (<i>n</i> = 2)	38.25 (19.2–57.3) (<i>n</i> = 1)	11.7 (0.0–25.9) (<i>n</i> = 4)
SODA	79 (75–83)	78 (64–83)	65 (51–79)	80	73.5 (57–84)
Total IPA	2.9 (1.0–4.8)	2 (1.6–2.8)	3.7 (1.8–5.5)	0.2	2.3 (0.4–5.6)
IPA problem perception	6 (5.0–7.0)	3 (0.0–3.0)	1 (1.0–1.0)	0	2 (0.0–7.0)

IPA, Impact on Autonomy and Participation Questionnaire; IQR, interquartile range; LRD, longitudinal radial deficiency; *n*, absolute number; *p*, significance; ROM, range of motion; SODA, Sequential Occupational Dexterity Assessment.

[‡]Patient with left type 2 B&K with operatively treated with centralization, right type 3 B&K operatively treated with opponensplasty. [‡]One of the patients also underwent a (unilateral) pollicisation.

lived their lives. They are not very keen on any further operation except when functional improvement is ensured.

Conservatively and operatively treated patients

No remarkable differences between conservatively and operatively treated patients were found. We realize that we only investigated centralisation as operative treatment in a very small study population, which might have influenced our results. Bora et al. [20] compared ten treated patients with three untreated patients. Their centralised and conservatively treated patients scored 54% and 27% of normal in active ranges of joint motion, respectively. Unfortunately, the patients were not assessed using objective tests to establish restrictions in activities. Further research is definitively needed to clarify this issue.

Correlation between function, activity and participation

Only a limited ROM of the fingers correlated with restrictions in activities. Buffart et al. [9] found a non-linear relationship between strength and activity and a more linear pattern between joint mobility and functional activities in children, which results are confirmed by our study. These results suggest that improving mobility could have a positive effect on performing activities but not on participation. Goldfarb et al. [19] alludes to this dilemma in their discussion regarding the lack of correlation between the results of the Jebsen test and the DASH questionnaire with body function or structure (i.e. forearm length, angulation of wrist, grip and pinch strength) in their study. Escalante et al. [25] studied how much disability is explained by RA in patients with this disease. They concluded that the level of disability was not proportional to the severity of patient's RA signs and symptoms. Their study confirmed the clinical impression that the manifestations of RA is the underlying factor determining the overall outcome. However, they also demonstrated the importance of demographic and psychosocial characteristics. The ICF also underlines the relevancy of personal factors and environmental factors in their framework [26]. The fact that the ability to perform activities and to participate in society is only influenced to a limited amount by restrictions in body functions is valuable information for parents of children with LRD. For them it can be reassuring to know soon after the diagnosis that children with severe deficiencies can fulfil societal roles in a satisfactory way later in their lives.

Limitations

This present study has limitations. The incidence of people with LRD is low; the results are therefore based on a small group of patients. Combined with the heterogeneity in the group (mild and severe types, bilaterally and unilaterally affected patients, operatively and conservatively treated patients), it was very difficult to compare these patients. However, the primary purpose of this study was to evaluate long-term activity and participation outcomes. As such, the heterogeneity did not weaken our conclusion that the severity of LRD does not correlate with more limitations. Twelve patients did have an associated syndrome. It could be that the associated syndrome had a negative influence on activity and

participation. To reduce this influence patients were given very clear instructions to focus only on the affected arm when judging their complaints and restrictions. More research is needed to identify people's participation level objectively so that outcomes can be better compared. Future research should use several measures of participation, more activity measures that include those commonly assessed during rehabilitation and functional measures that are scaled for difficulty in movement of hands and fingers [24].

In conclusion, patients with LRD, especially severe LRD, perform low in body function and structure tests and relatively high in participation and activity scores. The severity of body function structure in LRD deformities partially determines activity but does not determine participation in adult life.

Declaration of Interest: The authors report no declaration of interest.

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